Original Article

Clinical Presentation of Dilated Cardiomyopathy in Children

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ABSTRACT

Objective: To describe the presenting symptoms of dilated cardiomyopathy in children.
Study Design: Descriptive Case series.
Place and Duration of Study: This Study was conducted at the Department of Pediatrics, Bolan Medical Complex Hospital, Quetta during a period of 1 year.
Patients and Methods: Following patients were included in this study; that had tachycardia, respiratory distress, and heart failure. Following patients were excluded from this study; hypertrophic cardiomyopathy, restrictive cardiomyopathy, coronary artery anomalies, congenital heart disease, Rheumatic carditis and RHD. 50 Patients were included and their clinical variables, laboratory parameters and echocardiograms were analyzed.
Results: Out of 50 patients, 5 died, all these were 10 years or older. The following symptoms were found to be clinically significant. Respiratory distress $p<0.05$, poor feeding $p<0.05$. The signs were clinically significant. Arrhythmia $p<0.05$, Fever $p<0.05$, Tachycardia $p<0.05$, Tachypnea $p<0.05$, Hepatosplenomegaly $p< 0.05$, gallop rhythm $p<0.05$. Other significant findings were: X-ray chest, increased cardio thoracic ratio $p<0.05$. On echocardiography increased dimension >115 and ejection fraction <20% $p<0.05$, ST segment changes and T wave abnormality $p<0.05$.
Conclusion: High index of suspicion followed by appropriate investigation can lead to correct diagnosis.
Key words: Dilated cardiomyopathy, Echocardiography, Electrocardiography, Tachycardia

INTRODUCTION

Dilated cardiomyopathy in childhood is a diverse disorder with outcomes that depend on cause and age at presentation, as well as heart failure status.[1] The incidence of DCM is 0.56 cases per 100 000 per year, 10-fold lower than in adults. Boys have a higher DCM incidence than girls related to X-linked genetic causes and neuromuscular disorders. Black children have higher rates of DCM and different causes of DCM than do white children.[2] The majority of children who are diagnosed with dilated cardiomyopathy come to early medical attention because of severe symptoms.[3] Congestive heart failure was the initial symptom in almost 90% of patients, sudden death was the first manifestation of dilated cardiomyopathy in nearly 5%, and a further 13% died during their initial hospitalization.[4] In children with dilated cardiomyopathy, lymphocytic myocarditis is found more frequently and more commonly reflects a viral origin.[5] Other potential causes, such as familial dilated cardiomyopathy, a metabolic disease and parental consanguinity (as a marker for a recessively inherited condition), were each documented in 8.8% to 14.7% of study subjects. Familial cardiomyopathy and mitochondrial diseases may well have been under recognized, because not all subjects and families were systematically screened.[6] Echocardiography is an important tool to diagnose and differentiate dilated cardiomyopathy from other causes of cardiac failure.[7] Studies from 1975-1990 reported 70% survival at 2 years and 52% survival at 11.5 years of follow-up.[8,9] However, a publication from Texas that included patients diagnosed from 1990-2004, has reported a survival of only 40% at a mean follow up 6.2 years.[10]

PATIENTS AND METHODS

This study was carried out at Bolan Medical Complex Hospital Quetta, for a period of one year. During this time, 50 patients were clinically diagnosed dilated using non-probability convenience sampling. Patients under the age of 12 years who presented with respiratory distress, tachpnea, tachycardia, shock, increased capillary refill(>2 Sec), heart failure, ejection fraction of the left ventricle<45% on echocardiography, fractional shortening < 25% on echocardiography and/or left-ventricular end-diastolic diameter > 117% of the predicted value corrected for age and body surface area were included. Patients with congenital heart disease, pericardial disease, supraventricular tachycardia, systemic disease associated with dilated cardiomyopathy, hypertrophic cardiomyopathy, restrictive cardiomyopathy, rheumatic heart disease, coronary artery anomalies, cor pulmonale, and

Kawasaki disease were excluded. All patients who presented with the symptoms mentioned in inclusion criteria were subjected to detailed history, physical examination and Proforma was filled. Those who had any features mentioned in the exclusion criteria were dropped. All this data in the record file was subjected to analysis on SPSS version 10 and subsequently reported. P value was calculated for quantitative variables, Confidence interval for qualitative variables.

RESULTS

During one year the total number of patients admitted in our unit was 1240. Out of these the patients with cardiac disease were 20.48%. Out of these dilated cardiomyopathy was diagnosed in 19.68% of patients. The relative risk was 0.2 %(clinically insignificant). The period prevalence was 4.3 cases per 1000 population. The male to female ratio was 1:1.4 (95% CI 0.30-0.58) and female constituted 56% of the total no of patients. Almost 70% patients in the 1-5 years age group. The mean was 5.74± 0.59, median was 3.75 and mode was 2. The standard deviation is 4.18; variance 1748, range is minimum 1 and maximum 12. Almost 80%of the patients presented within the first week of their illness, and only 6% presented in the third week(95% CI 1-4 wks) with the remaining presenting in the second week(Chi sq 39.600, df 13, asymp sig.000). 60% presented in grade 2 severity according to New York Heart Association (NYHA) classification (95% CI 49-64%), with 14 % presenting in grade 3 (95% CI 33-44%), and 6%(95% CI 5-8%) in grade 4 and 20% (95% CI 15-22%) in grade 1 respectively. The chi sq is 31.600, df 3, and asymp sig .000. The frequency of symptoms at presentation is given in table No.1.

The percentage of signs in DCM is given in table 2. Chest X-ray were taken in all patients, the most common finding was cardiomegaly, increase vascular marking were present in 50% of the patients (95% CI 44-52%) (Chi sq 0.000, df 1, asymp sig 1.000), co-existent pneumonia was present in 38% (95% CI 37-40%), while pleural effusion was found in only 10% (95% CI 8-12%).

The echocardiographic findings are given in table 3 along with percentages and CI. Followed by ECG findings and percentages in table 4.

Total leucocyte count was normal in 48% cases(95% CI 4000-11000), it was increased in 52%(95% CI 11000-18000), while it increased above 20,000/cm only in 26% of patients (20500-50000).64% 0f patients having Hb% below 10g/dl(95% CI 5-8),24% were significantly anemic with Hb% below 4 gm/dl(95% CI 2-5%) while the rest had normal Hb. Serum Electrolytes were done in all cases with hyperkalemia present in 36%(95% CI 5.0-6.5 mEq/dl) and hypokalemia in 30%(95% CI 2.5-4.5 mEq/dl),while the rest had normal K levels. 10% of the patients expired, the rest were discharged.

Table No.1: Frequency and percentage of symptoms at presentation

Symptoms	Frequency	Percentage
Cough	32	64.0
Respiratory distress	30	60.0
Poor feeding	25	50.0
Irritability	20	40.0
Exercise intolerance	10	20.0
Abdominal pain	8	16.0
Poor weight gain	5	10.0

Table No.2: Percentage of signs in DCM

Sign	Frequency	Percentage
Tachypnoea	50	100.0
Tachycardia	50	100.0
Gallop rhythm	43	86.0
Increased liver size	42	85.0
Chest indrawing	20	40.0
Pallor	15	30.0
Murmur	15	30.0
Pulmonary edema	15	30.0
Decreased capillary refill	10	20.0
Orthopnea	10	20.0
Increased JVP	8	16.0
Vomiting	5	10.0
Irregular pulse	5	10.0

Table No.3: Echocardiographic findings with frequency and CI

Findings	Frequency	CI
Chamber dimensions increased	100%	95%-105%
Ejection fraction decreased	100%	95%-105%
Regional wall abnormality	54%	55%-61%
Mitral regurgitation	30%	37-44%
Tricuspid abnormality	20%	18-24%
Mural thrombus	4%	1-4%

Table No. 4: ECG Findings along percentages and CI

Findings	Percentage	CI
T wave abnormalities	70%	65%-74%
ST segment abnormalities	64%	62%-69%
P wave abnormalities	16%	
Left bundle branch block	10%	7%-12%
Atrial fibrillation	6%	4%-7%

DISCUSSION

The prevalence of DCM in Bolan Medical Complex Hospital, Quetta was found to be 19.68% while at

AKUH it was 13% of all cardiac disease.[11] The latter study extended over 7 years period, similarly Kothari from New Delhi founded 145 children over a period of 9 years.[12] This percentage is higher significantly and calls for the need for better understanding and early diagnosis through conducting further studies .

Our study had a female preponderance that is 56% females, 44% males and male to female ratio was 1:1.5. Shabina Ariff reported male predominance male to female ratio 1:0.6. Similarly Kothari from India reported a male predominance 1.8:1; however Elnoor has reported a female predominance, with male to female ratio 1:3.5, Nonogueria from Portugal studies 34 children with cardiomyopathy with male to female ratio 1:4. Now whether these reports from different geographical areas are coincidental or represent unknown factors is not known.

Our study found that age group most strongly affected 1-5 years age groups. This is consistent with other studies. Agarwall et al from Muscat showed that 83% of children were below 2 years.[13] Similarly Taliercio et al also showed the preponderance of infants and younger children.[14] Regional data shows similar results. Dalal in India in hospital based study showed 20 children with dilated cardiomyopathy that presented most frequently between 2-5 years. A Finish study had identical results with all patients presenting below 2 years of age.

In our study 94% of the patients presented with in the first 2 weeks of onset of illness. This is not consistent with National Data. Liaqat Ali in his land mark study showed that 33% patients presented in first 6 months.[15] But this was a study of adult patients. Kothari showed that the mean duration illness at presentation was 5.8 months, this was found to be statistically insignificant ($p > 0.05$)

The severity of symptoms were judged by New York Heart association classification, while we found only 30% of patients in class ¾, Kothari demonstrated 38% patients. Most of the patients in our study presented in class I and II. This was found statistically insignificant ($p>0.05$)

The following symptoms were found to be statistically significant. Respiratory distress ($p<0.05$), poor feeding ($p<0.05$), cough ($p<0.05$). The following signs were found to be statistically significant: Arrhythmia, fever, Hepatomegaly, tachycardia, tachpnea, gallop rhythm($p<0.05$). Cardio thoracic ratio more than 55% was found to be statistically significant (<0.05).

The following finding on Echocardiography were found to be statistically significant ($p<0.05$). Decreased left ventricular ejection fraction, increase chamber dimension and regional wall abnormality.On ECG ST Changes and T wave abnormality, left ventricular hypertrophy were found to be statistically significant (>0.05).

This study has high mortality rate that is 10%, it would be pertinent at this point to list the factors found on univariate analysis to be statistically significant.

The higher age of diagnosis higher cardiothoracic ratio > 60% and higher ratio of LVED/PW thickness were associated with higher mortality rate. Similarly patients with Hemiplegia mural thrombus, very low ejection fraction of less than 20% arrhythmia, hyperkalemia.

The outcome of this study was better than other studies conducted nationally and internationally e.g. Shabina Ariff reported 28% mortality, while studies conducted in America, Australia, Japan, Finland, Africa, India have reported higher mortality rates as high as 60%.

This difference was probably due to lack of follow up of our patients. Our consideration was single admission and its outcome that was either discharge or death, perhaps many of the children died latter or referred to other tertiary care centers. While compared to the meager resources at our disposal, the mortality is still surprising low. For example Shabina Ariff reported a mortality of 28% at AKUH that is the most sophisticated specialized center in our country and this was during a course of single admission.

A Kagi T from Hospital for sick children Toronto reported a similar outcome; despite intensive medical therapy DCM in children is associated with survival rate of 41% at one year and 20% at 3 years after diagnosis. In the same vein Ghani VK and Colleagues from Ahmed Abad India showed that despite vigorous therapy course was rapidly downhill and prognosis poor, similarly data from Italy showed that only 38.2% children survived at the end of single admission, while mortality was 29%. The Finish study concluded that children with DCM had a mortality of 50% and after 5 years only 20% were alive, it also showed that male patients less than 1 years of age had poor outcome compared to female of the same age group (p0.09)

On the other hand our study showed that all five deaths were among male patients and they were all more than 10 years of age. Shabina Ariff also showed that the mortality in male is two and half time more than females. Hence in the two Pakistani studies male gender had the significant impact on the outcome whether this is actually the case is controversial as results from different studies are conflicting. Another interesting feature of this study is that highest mortality was seen in children 10 years or above.

This is consistent with National and international data e.g. Shabina Ariff showed a mortality of 50% in the older age group, similarly Agarwall et all showed that the outcome was poor in older age group; in contrast Taliercio et al showed that children under the age of 2 years had a higher fatality rates of 90% over 2 years. Griffin on the other had showed a poor outcome for children more than 2 years of age.[16] Dalal in India did not observe any difference in outcome and age.

In our setup it could be due to lack of differentiation of cardiomyopathy as a case of respiratory distress, low threshold for referral for young children with presumed pneumonia, Who classification for pneumonia that has resulted in ambiguity among primary physicians alike regarding cardiac etiologies causing respiratory insufficiency.

Alternatively this could be due to different causes of cardiomyopathy in different age groups, relatively immature cardiac system and myocardial metabolism of the infant, limited adoptive mechanism stress that could lead to severe yet reversible myocardial dysfunction perhaps the regenerative capacity of infant myocytes enhanced hypertrophic tendency in response to wall stress and less after load secondary complaint peripheral artery . Many have postulated that children less than two years of age who are diagnosed as having DCM may actually have a slowly resolving myocarditis for resolving this grey area. Further studies are needed focusing on specific age groups, their etiology and outcome to accurately point out true risk age group. Our study did not encounter any positive family history in all 50 patients included. Arrhythmias and mural thrombosis which were found in a very small percentage of patients were associated with Hemiplegia and death, since the size of sample is too small , whether a temporal relation exist cannot be commented upon. In conclusion it would be safe to say that we lack data regarding etiology, risk factors, and outcome on DCM in our part of the world.

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